WHAT IS CLAIMED IS:

1. An isolated CLN2 protein with the following characteristics:

a) said CLN2 is a protein with pepstatin-insensitive carboxyl protease activity; and,

b) mutation or absence of said CLN2 is causative of classical late infantile neuronal ceroid lipofuscinosis (LINCL).

2. A chimeric protein comprising the CLN2 protein of Claim 1.

3. A purified nucleic acid encoding CLN2, or a fragment thereof having at least 15 nucleotides.

4. The nucleic acid of Claim 3 which encodes CLN2 having an amino acid sequence as depicted in Figure 3 (SEQ ID NO:3).

5. The nucleic acid of Claim 4 having a nucleotide sequence as depicted in Figure 3 (SEQ ID NO:1), corresponding allelic genes, homologous genes from other species, and nucleotide sequences comprising all or portions of *CLN2* genes which are altered by the substitution of different codons that encode the same amino acid residue within the amino acid sequence (SEQ ID NO:3), thus producing a silent change.

6. The purified nucleic acid of Claim 3 which is DNA.

7. A recombinant DNA expression vector comprising the DNA of Claim 6, wherein the DNA encoding the CLN2 is operatively associated with an expression control sequence.

8. A transformed host cell comprising the DNA vector of Claim 7.

9. A recombinant virus comprising the DNA vector of Claim 7.

10. The recombinant virus of Claim 9 selected from the group consisting of a retrovirus, herpes simplex virus (HSV), papillomavirus, Epstein Barr virus (EBV), adenovirus, and adeno-associated virus (AAV).

11. A method for producing a CLN2 comprising culturing the transformed host cell of Claim 8 under conditions that provide for expression of the CLN2.

12. The method according to Claim 11 wherein the host cell is a bacterium.

13. The method according to Claim 11 wherein the host cell is a mammalian cell.

14. A method for increasing the level of expression of a CLN2 comprising introducing an expression vector of Claim 7 into a host *in vivo* under conditions that provide for expression of the CLN2.

15. The method according to Claim 14 wherein the expression vector is a viral expression vector.

16. The method according to Claim 14 wherein the expression vector is a naked DNA expression vector.

17. A method for treating LINCL in an animal by increasing the level of CLN2 in cells.

18. The method according to Claim 17, wherein the level of CLN2 is increased by administration of CLN2 to the animal.

19. The method according to Claim 18, wherein the level of CLN2 is increased by administration of a recombinant expression vector to the affected cells, which expression vector provides for expression of the CLN2 *in vivo*.

20. An oligonucleotide of greater than 20 nucleotides which hybridizes under stringent conditions to the nucleic acid of Claim 3, wherein the T_m is greater than 60°C.

21. The oligonucleotide of Claim 20 which is an anti-sense oligonucleotide.

22. An antibody specific for CLN2 of Claim 1.

23. The antibody of Claim 22 which is labeled.

24. A method for detecting CLN2 in a biological sample comprising:

a) ontacting a biological sample with an antibody of Claim 22 under conditions that allow for antibody binding to antigen; and,

b) detecting formation of reaction complexes comprising the antibody and CLN2 in the sample;

wherein detection of formation of reaction complexes indicates the presence of CLN2 in the sample.

25. A method for quantitating the level of CLN2 in a biological sample comprising:

a) detecting the formation of reaction complexes in a biological sample according to the method of Claim 24; and,

b) evaluating the amount of reaction complexes formed;

wherein the amount of reaction complexes corresponds to the level of CLN2 in the biological sample.

26. A method for measuring CLN2 activity in a biological sample based on the amount of CLN2 pepstatin-insensitive carboxyl protease activity relative to normal controls in a biological sample.

27. A method for detecting CLN2 in a biological sample comprising:

a) contacting a biological sample with an oligonucleotide of Claim 21 under conditions that allow for hybridization with mRNA; and,

b) detecting hybridization of the oligonucleotide to mRNA in the sample;

wherein detection of hybridization indicates the presence of CLN2 in the sample.

28. A method for quantitating the level of CLN2 in a biological sample comprising evaluating the quantity of oligonucleotide hybridized according to the method of Claim 26, wherein the quantity of oligonucleotide hybridized corresponds to the level of CLN2 in the biological sample.

29. A method for detecting the *CLN2* gene, and mutant variants associated with LINCL, in chromosomal samples comprising of:

a) contacting a chromosomal sample from, for example, amniotic fluid, with an oligonucleotide of Claim 21, or variants of said oligonucleotide that hybridize to mutant alleles of *CLN2*, under conditions that allow for hybridization; and,

b) detecting hybridization of the oligonucleotide to the chromosomes in the sample;

wherein detection of hybridization is used a method of prenatal screening for LINCL.

30. A method for identification of lysosomal proteins based on the presence of mannose 6-phosphate glycosylation and comprising the following steps:

a) purifying proteins from a biological sample using an affinity column consisting of the mannose 6-phosphate receptor immobilized on a solid support;

b) peptide sequencing of selected purified proteins;

c) designing nucleic acid probes based on the peptide sequences derived in step b; and,

d) using the probes of step c to isolate and characterize the genes encoding the purified lysosomal proteins.